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Case Report

A diagnostic challenge in a child: a rare brain tumour or tuberculous meningitis

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ABSTRACT

A case of non-responding tuberculous meningitis in a 10-year-old male child who was diagnosed and received Anti-Tubercular Therapy twice. After exploring for alternative diagnosis, biopsy was suggestive of DLGNT. High prevalence of tuberculosis in India and similarity in MRI brain findings might have contributed for delay in diagnosis. Leptomeningeal enhancement in Paediatric patients is a common MRI finding associated with numerous conditions including infections, inflammatory process, neoplasms, and post-ictal hyperaemia. Presence of leptomeningeal enhancement along with hydrocephalous in the patients living in developing world are often labelled as tuberculous meningitis. DLGNT is a rare neoplasm, which was previously known as a disseminated oligodendroglia-like leptomeningeal tumour of childhood. DLGNT has been defined as a neuronal/glioneuronal tumour as per the 2021 World Health Organization (WHO) classification of brain tumours. This tumour may also present with hydrocephalous in initial stages. The confirmatory diagnosis of tuberculous meningitis is often difficult as its clinical features are not very specific. Detection of Mycobacterium tuberculosis in cerebrospinal fluid (CSF) by acid-fast staining, culture, or DNA analysis with polymerase chain reaction (PCR) has low sensitivity. DLGNT needs histologic confirmation through brain biopsy. Although rare, DLGNT should be considered as a differential diagnosis. Especially in children who are solely diagnosed on radiological evidences of tuberculosis. The current case highlights the importance of histologic confirmation through brain biopsy for cases presenting leptomeningeal enhancement in the basal cistern in MRI with equivocal laboratory examinations to explain the aetiology.

Keywords: Tuberculous meningitis, DLGNT, Neuronal/glioneuronal tumour

INTRODUCTION

Leptomeningeal enhancement in paediatric patients is a common MRI finding associated with numerous conditions including infections, inflammatory process, neoplasms, and post-ictal hyperaemia. Presence of leptomeningeal enhancement along with hydrocephalous in the patients living in developing world are often labelled as tuberculous meningitis. DLGNT is a rare neoplasm, which was previously known as a disseminated oligodendroglia-like leptomeningeal tumour of childhood. DLGNT has been defined as a neuronal/glioneuronal tumour as per the 2021 WHO classification of brain tumours.[1] This tumour may also present with hydrocephalous in initial stages. The confirmatory diagnosis of tuberculous meningitis is often difficult as its clinical features are not very specific. Detection of *Mycobacterium tuberculosis* in CSF by acid-fast staining, culture, or DNA analysis with PCR has low sensitivity.[2] DLGNT needs histologic confirmation through brain biopsy.

CASE REPORT

A 7-year-old male child had one episode of generalized convulsion associated with fever in 2018. He was completely immunized as per National immunisation schedule with no significant birth or family history. He was developmentally normal and anthropometric measurements were within expected norms. He had signs of meningeal irritation at admission.

Neuroimaging showed leptomeningeal enhancement, Communicating Hydrocephalus with periventricular oedema. Ventriculoperitoneal Shunt was placed and category 1 antituberculosis therapy was started along with steroids and antiepileptic drugs. He completed Anti-tuberculosis therapy for 1 year. His compliance was good and was asymptomatic until 2021.

In October 2022, patient was admitted for drowsiness, one episode of convulsion and altered sensorium. MRI done, which showed Diffuse leptomeningeal enhancement with VP shunt in situ. Most likely differential diagnosis was Diffuse leptomeningeal glioneuronal tumour which required histopathological confirmation. Patient was referred to the neurosurgery department for biopsy.

Suboccipital craniotomy and biopsy of left cerebellar hemisphere lesions was done, under intraoperative ultrasonographic guidance. Histopathological examination was consistent with low grade glial / glioneuronal tumour. After diagnosis, he was started on targeted chemotherapy. Patient is on close follow up and he did not show any worsening of symptoms.

Imaging findings

MRI showed cystic-like lesions on T1-weighted images, hyperintense lesions on T2-weighted images, with no restriction on diffusion-weighted imaging and minimal leptomeningeal enhancement on post-contrast images as shown in Figure 1.

MRI findings of DL-GNT are diffuse well defined no enhancing cystic lesions appearing hypointense on T1 W images (a) and hyperintense on T2 W images (b) involving leptomeningeal surface of bilateral cerebellar hemisphere and temporal lobe with minimal suppression on long TR FLAIR images.

These lesions show no restriction DW images (e) and does not bloom on GRE images (d). Coronal T2 W image (f) shows dilated ventricles with VP shunt in right lateral ventricle with surrounding mild periventricular oedema. Coronal post contrast T1 fat supressed image (g) shows minimal leptomeningeal enhancement along bilateral cerebellar hemispheres. (Figure 3 a-g). The Biopsy showed the cells within leptomeninges which were compost of oligodendrocytes with increased mitotic activity, immunopositivity for Olig2 and S-100.

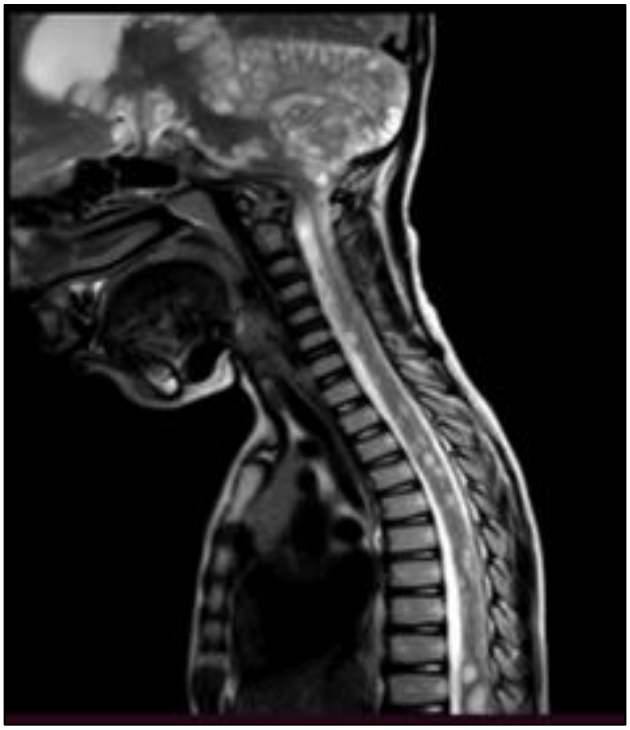

Figure 1: Sagittal T2-weighted image of the cervicothoracic spine shows cystic lesions involving brain and spinal cord.

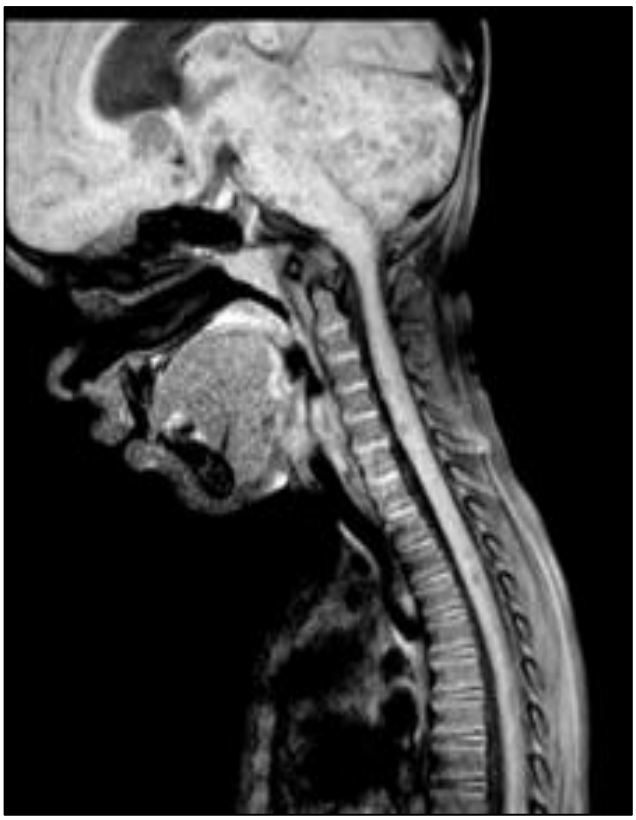

Figure 2: The cystic lesions do not enhance on post contrast studies.

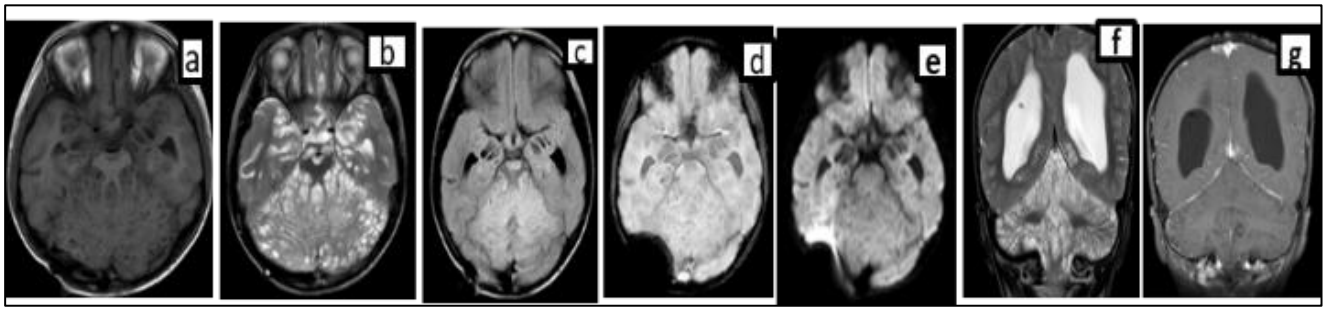

Figure 3 (a-g): MRI findings of DL-GNT.

DISCUSSION

This case emphasizes the diagnostic challenges faced in identifying DLGNT in paediatric patients. The initial presentation of seizures and hydrocephalus led to a misdiagnosis of Tuberculous meningitis (TBM), as DLGNT shares similar clinical and radiological features, delaying the correct diagnosis. In this case, despite of treatment for TB Meningitis, the symptoms persisted, prompting further investigations and ultimately leading to the correct diagnosis.

In cases where standard therapies do not yield improvement, it is crucial to reconsider the primary diagnosis. Similar cases have been reported by Schwetye et al and Lee et al.[3-5] The correct identification of DLGNT enabled the initiation of targeted chemotherapy, highlighting the importance to distinguish between infectious and neoplastic causes, for proper management.

DLGNT has been defined as a Diffuse Glioneuronal Tumour as per the 2021 World Health Organization WHO classification of brain tumours.[1] Usually, the age at diagnosis is 8 years, with a male predominance of approximately 2:1. DLGNT was classified as a distinct entity in the 2016 WHO classification of brain tumours.[1] It primarily affects male children, with a very rare occurrence in adults.[1]

Histologically, DL-GNT is characterized by the widespread proliferation of oligodendroglia-like cells, without infiltrating the brain parenchyma.[2] DLGNT is classified into low grade and high grade based on molecular and histopathological features. Paediatric DL-GNT is typically a slow-growing tumour with a low proliferation rate, as indicated by Ki-67 immunohistochemistry. In low-grade cases, the average survival is 22 months, with 63% of patients passing away within 9 months of diagnosis.[3] However, high-grade DL-GNT in children is rare and typically follows a more aggressive clinical course. The exact incidence of this condition remains unclear. This can be due to the paucity of information of few individual case reports and small case series.

CONCLUSION

Although rare, DLGNT should be considered as a differential diagnosis. Especially in children who are solely diagnosed on radiological evidences of tuberculosis. The current case highlights the importance of histologic confirmation through brain biopsy for cases presenting leptomeningeal enhancement in the basal cistern in MRI with equivocal laboratory examinations to explain the aetiology.

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